

# Turgenev's 'living relic': an early description of scleroderma?

Richard M Ellis FRCS FRCP<sup>1</sup> Rupak Moitra MRCP<sup>2</sup> Nigel North PhD<sup>3</sup>

*J R Soc Med* 2005;98:372–374

Can a lay person accurately report the external features of disease, or is a medical training necessary? Turgenev's short story *Living Relic*, published in 1874, gives a detailed description of a young woman afflicted by a debilitating, chronic illness.<sup>1</sup> The story is told by Pyotr Petrovich Karataev, the son of a landowning family who has the leisure to enjoy hunting and shooting, who when visiting one of his mother's farms, stumbles on a wattle shed in the bee garden: to his surprise this deserted spot reveals a woman lying on a simple bed of boards, under a quilt.

'The head was completely withered, of a uniform shade of bronze, exactly resembling the colour of an ancient icon painting; the nose was as thin as a knife-blade; the lips had almost disappeared—only the teeth and eyes gave any gleam of light... At the chin, where the quilt was folded back, two tiny hands of the same bronze colour slowly moved their fingers up and down like little sticks. And the face seemed all the more awesome to me because I could see that a smile was striving to appear on it, to cross its metallic cheeks—was striving and yet could not spread.'

With a shock, after the woman had said, "'Master, don't you recognize me?'"... he realizes that she is Lukeria, who ten years before had been 'the greatest beauty among all the servants in our house, tall, buxom, white-skinned and rosy-cheeked, who used to laugh and sing and dance!'

She tells him to come and sit close to her, after which she 'spoke very quietly and faintly', and she recalls how six or seven years ago (she is now 29) her illness had started, apparently, when she got up at night, not being able to sleep, and slipped off the porch and 'flew down—bang—onto the ground... it was just like something inside—in my stomach—had broken... Ever since, I began to wither and sicken, and a blackness came over me and it grew difficult for me to walk and I even began to lose control of my legs. I didn't feel like eating or drinking.' Currently she

survives '“So far as feeding goes, I don't eat anything, and I have water—there it is in that mug... I can stretch out, myself, because I've still got the use of one arm.”' Elsewhere she describes musculoskeletal symptoms in further detail

“‘Not a single one of the medicals could even say what kind of an illness I had... Once a doctor came... He started turning me this way and that, straightening and bending my legs and arms... for a whole week afterwards my poor bones were aching’... ‘I may not have big pains, but something is always gnawing at me right there inside me, and in my bones as well.’”

Lukeria 'sighed with difficulty. Like other parts of her body her breast would not obey her wishes.' But she says, "'Don't you pity me too much... I sing songs even now.'"

## THE SOMATIC DISEASE

Turgenev provides fine detail in his description of the woman, the overall impression being of smallness and her head 'withered'. Clearly with her stomach problems and almost non-existent diet she must have lost weight to an extreme degree, but two striking aspects are the bronze pigmentation of the face (and hands) and its mummy-like immobility. In addition the mouth was much reduced in size and the nose thin. The third major observation was the stiffness of her joints. This was combined with the finger deformities 'like sticks'.

Our clues to the diagnosis are skin pigmentation, a mask-like face with almost no lips, deformities of the fingers with general stiffness and pain in the limbs. This is combined with a systemic illness with abdominal pain and an inability to take more than a liquid diet. From the start we considered scleroderma.

The pigmentation raises possibilities such as haemochromatosis, but, even with liver failure and diabetes, abdominal pain and facial deformity would not have occurred, and the joint involvement of haemochromatosis does not produce stick-like fingers even if the metacarpophalangeal joints are affected. Addison's disease, which tends to cause pigmentation on the trunk and pressure

<sup>1</sup>University of Southampton; <sup>2</sup>North Hampshire Hospital, Basingstoke;

<sup>3</sup>Salisbury District Hospital, Salisbury, UK

Correspondence to: Dr R M Ellis, Rheumatology, Mailpoint 78, General Hospital, Southampton SO16 6YD, UK

E-mail: UKjom2@aol.com

areas, likewise does not fit. Prolonged malabsorption (for example from Crohn's disease) would not explain the lips, and this type of hand deformity is again unlikely even if some peripheral arthritis occurs.

Other conditions associated with the appearance of thickened 'sclerodermatous' skin must be considered. Widespread scarring and dermal fibrosis has been reported after injury to the spine and central nervous system. While the possibility of a traumatic precipitant certainly exists in the case of Lukeria, the extracutaneous features and characteristic peripheral stigmata are at odds with this interpretation. The same applies to alternative causes of skin thickening such as scleromyxoedema, sclerosing panniculitis and acrodermatitis chronica atrophicans. The combination of thickened skin, hyperpigmentation, neuropathy and weakness is occasionally seen in amyloidosis and also in certain types of porphyria and the plasma cell dyscrasia POEMS (polyneuropathy, organomegaly endocrinopathy, M protein and skin changes); but the age of onset of Lukeria's symptoms and their progressive nature make these diagnoses unlikely.

We have come to the view that Lukeria's features are all consistent with scleroderma (systemic sclerosis) of the diffuse type. Although onset is most commonly in the fourth and fifth decades of life, the disease can start in the second and third decades: 8% were under age 24 at first diagnosis in a series from a county hospital in the USA.<sup>2</sup> The facial skin change produces an appearance which is 'pinched immobile and expressionless'<sup>3</sup> and the lips are thin with a characteristic narrowing of the mouth which interferes with eating. Skin pigmentation can increase or decrease, and even though in the late phases the skin can soften, it often remains pigmented. Involvement of the gastrointestinal tract is considered the second most frequent feature in scleroderma after the skin and is more common than joint disease. D'Angelo confirmed this with his combined necropsy and clinical study.<sup>4</sup> Recent work showing impaired blood flow to the small intestine, even in patients without gastrointestinal symptoms, is further evidence of the frequency of gut involvement.<sup>5</sup> The vascular changes of fibrosis and arteriosclerosis are widespread in visceral organs,<sup>4</sup> leading to weight loss and often profound fatigue. The resultant disorder of motility and bacterial overgrowth contribute to the malabsorption. Dysphagia typically causes patients to reduce food intake and to avoid solids. In the lower oesophagus the common lesion is disorganized peristalsis and relaxation of the gastro-oesophageal sphincter, leading to gastro-oesophageal reflux and oesophagitis. Skin involvement in the hands, with initial puffiness and loss of skin creases, leads to later soft tissue absorption especially of the distal finger pulp, with or without vasculitic lesions, and the associated immobility can give the 'stick-like' fingers of sclerodactyly. In addition, as

originally researched by Rodnan,<sup>6</sup> there is a true erosive arthropathy in 20–30% of cases<sup>7</sup> resulting from an inflammatory synovitis without pannus but with an intense fibrosis and narrowing of the joint space. Lukeria's leg weakness is difficult to ascribe definitely to a primary muscular, neurological or arthritic cause. In scleroderma, muscle atrophy is most often the result of disuse, with joint involvement, but can be due to a true myositis.

## PSYCHOLOGICAL ASPECTS

Turgenev's description of Lukeria's physical plight is striking but so also is his description of her ways of coping with her handicaps, pain and progressive debility. We have therefore analysed *Living Relic* from a psychological standpoint. In the story we are introduced to a young woman, formerly much admired and a beautiful dancer who now has a chronic debilitating condition.

It is noteworthy that despite the chronicity and disabling nature of her condition, Lukeria displays none of the expected behavioural features.<sup>8</sup> Indeed, the narrator is amazed by 'the almost gay manner in which she was telling her story, without groans or sighs, never for a moment complaining or inviting sympathy.' She gets enjoyment from helping the orphan girl to learn singing: "'I've been teaching her and she's picked up four songs already.'" She uses such distractions to cope with her disability. "'Yes, I sing songs, the old songs, roundelays, feast songs, holy songs, all kinds! I used to know many of them after all; and I haven't forgotten them.'" She demonstrates a strong internal locus of control<sup>10</sup> as well as valuing interaction with other people. She tells us that 'people must help themselves'. Lukeria also uses reverie as a form of coping:

'"'Sometimes I lie by myself like I am now—and it's just as if there was no one on the whole earth except me. And I'm the only living person! And a wondrous feeling comes over me, as if I'd been visited by some thought that seizes hold of me—something wonderful it is. . . . It comes out like a cloud and pours its rain through me, making everything so fresh and good.'"

This is an example of 'dissociation'—a term that embraces minor events such as day-dreaming through to more extreme states where individuals 'remove' themselves mentally as a coping mechanism. Dissociation can be a positive experience for individuals coping with trauma or other distress. Dissociation permits the isolation of a traumatic experience until the individual feels able to cope with it.<sup>12</sup>

Thus, in Lukeria, we see all the positive attributes of someone who is able to interact with the world. We see that she does not make fruitless catastrophic comparisons

with her former self. She does not dwell on her disability but compares it favourably to the pain and suffering of other people.<sup>8</sup>

## DISCUSSION

Although *Living Relic* was not published until 1874 (to raise funds in a year of famine in Russia), it almost certainly relates to the times described in *A Sportsman's Sketches* published between 1847 and 1852, and therefore before the emancipation of the serfs.

Turgenev, born in 1818 and brought up on his mother's estate at Spasskoye, south of Moscow, was aware of the injustices of life for the serfs under the Tsarist regime. His experiences at Berlin University made him an advocate of progress in the Western European mode.

We have presented Turgenev's Lukeria as a likely case of scleroderma/systemic sclerosis. We do not know that Turgenev had any special interest in medical matters (although he is certainly able to laugh at doctors' expense in this tale), or of any acquaintance of his who had this type of illness. But his description is so precise and life-like that we feel it must have been based on someone he knew. Scleroderma had been described at the probable time of writing (1850–1870)—especially in French publications<sup>13–15</sup> although the earliest known description is Italian.<sup>16</sup> The systemic nature of the diffuse form of the disease was not recognized until the 20th century.<sup>17</sup>

While Turgenev's stories are much loved because of their intriguing descriptions of his characters' personalities and their interactions with one another, his *Sportsman's Sketches* threw into relief the plight of the unenfranchized serfs of Russia at that date and what westernized minds saw as appalling social injustice. Turgenev was consequently regarded as subversive and was effectively exiled to France. In *Living Relic*, however, Lukeria's plight results not from social injustice but the vagaries of nature; indeed her 'owner' (Pyotr's mother) arranges for her to see a doctor and offers hospital treatment. Turgenev's epilogue for the story, whether the suffering was manmade or natural, was

taken from his fellow-Russian poet Tyutchev's stanza: 'Homeland of suffering: thou art the land of Russia'.

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